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The Main Problems of Parents of a Child With Epidermolysis Bullosa

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Epidermolysis Bullosa (EB) is a rare genetic blistering-skin disorder with varying degrees of severity, ranging from mild forms to severe forms, with chronic progression. The aim of this study was to identify and specify the problems of parents of a child with EB. Qualitative research methodology was used, comprising a series of semistructured interviews with eleven families. The key problems of parents were broken down into three themes, related to the child, the family, and the care providers. These themes comprised nine categories, including (1) the child being different, (2) the child suffering pain, (3) feelings of uncertainty, (4) restrictions on employment and leisure time, (5) difficulties in organization of care, (6) never being off-duty, (7) family problems, (8) ignorance and lack of skills of care providers, and (9) resistance to difficult care. Despite the great variance in clinical pictures of the different (sub)types of EB, the main problems parents experienced appear quite similar. However, the problems did appear to differ in extensiveness, intensity, and gravity.

Keywords: *chronic illness; illness and disease; parenting; children; qualitative research; skin disorders*

Epidermolysis bullosa (EB) is a rare skin disorder with a lifetime exceptional liability of the skin to blister after minor mechanical trauma. Because of a genetic abnormality, the different skin layers do not stick together adequately, so that the skin surface can easily break down after hitting oneself, rubbing, scratching, or swallowing. There are varying degrees of severity, ranging from mild forms with only local involvement of the skin surface and with no impact on overall longevity, to severe forms involving various organs with early postnatal death or chronic progression (Fine, Bauer, McGuire, & Moshell, 1999).

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According to the consensus classification, three major types of EB can be distinguished (please see the Appendix for details on EB abbreviations): (1) EB simplex (EBS), (2) junctional EB (JEB), and (3) dystrophic EB (DEB; Fine et al., 2000). Each main type consists of diverse subtypes with varying degrees of severity. For example, the children with the Weber-Cockayne and Köbner subtype of EB simplex are, in comparison with those with the Dowling-Meara subtype of EB simplex, relatively mildly affected. That is, children with the Dowling-Meara subtype present widespread blistering at birth, which gradually improves as the child grows older, whereas the Weber-Cockayne and Köbner subtypes show only mild blistering without lifetime improvement. From the second main type, junctional EB, the non-Herlitz subtype is also a relatively mild form, whereas most children with the Herlitz subtype die within their first two years because of failure to thrive or progressive respiratory obstruction. The third main type, dystrophic EB, is divided into two subtypes: dominant dystrophic EB (DDEB) and recessive dystrophic EB (RDEB). Children with mutilating recessive dystrophic EB

(RDEB-mut) are affected with the most severe form of EB, in which permanent scarring leads to mitten deformities of the hands and feet. The dystrophic EB subtype is, when compared with RDEB, a relatively mild form of EB, which allows for an almost normal quality and length of life.

Even so, children with all types of EB can be confronted with extensive and complex problems such as never-ending itch, repeatedly painful wound care, disfigurement, embarrassment, and difficulties in attaining social demands (Fine, Johnson, Weiner, & Suchindran, 2003; Hall, 2004; Horn & Tidman, 2002). The skin disease can also put immense demands on the caregiving parents (Fine, Johnson, Weiner, & Suchindran, 2005; Landsdown, Atherton, Dale, Sproston, & Lloyd, 1986; Lin & Caldwell-Brown, 1996). Landsdown et al. (1986) were the first investigators to focus attention on the practical and psychological problems of caregiving parents, such as difficulty with dressings or in obtaining suitable shoes, and the environment not being fully understanding or sympathetic. Ten years later, Lin and Caldwell-Brown (1996) emphasized psychological problems such as feelings of guilt and fear, because parents of children with EB have to deal with a serious congenital disease and its often-unknown illnesses. Most recently, Fine et al. (2005) investigated the impact a child with EB had on the parents' family size and relationship. They found that most parents chose not to have additional children because of this serious illness and the ensuing lack of energy they had for participating in activities as a couple.

The aim of this study was to gain a more in-depth understanding of the wide-ranging and complex problems parents of children with EB have to deal with. This aim included two questions: (1) What problems do parents of a child with EB actually experience as the most difficult ones? and (2) Are there differences in experiences between parents of mildly and severely affected children? To address these two questions, we adopted a qualitative research design in the form of semistructured interviews with parents of children with all types of EB.

Methods

Design

A qualitative research design was used in the form of semistructured interviews. The semistructured interviews made it possible to create an in-depth

understanding of the problems encountered by parents of children with EB. An interview guide was developed through an extensive document analysis of EB. The interview guide was subsequently pilot-tested on parents of a young child with EB. Thereafter, some interview topics and leading questions were refined. The final interview guide consisted of five themes focusing on (1) the family history, (2) daily activities and practical problems, (3) the impact of EB on the family and social life, (4) problems with treatment and support, and (5) future concerns.

Participants

The participants were recruited from the EB dermatology database of a university medical center in the Netherlands, which consisted of eighty two children from 0 to 18 years of age with EB. Since 1992, an EB team has been available at the medical center that functions as a center of expertise for the Netherlands and Belgium. To date, more than 250 patients with EB have consulted this patient service. The EB team aims at early diagnosis and optimal and concerted management of the patients. The EB team employs interdisciplinary practice, thus providing superior patient care by combining the unique expertise of all twenty professionals, who are coordinated by a nurse practitioner. This results in high-level, demand-driven, coordinated, and collaborative care for EB patients. In addition, the EB team is in close contact with a patient organization which supports a social worker in the home setting of the patients.

Parents who met the following criteria were included in the study: those who (1) were raising a child diagnosed with a (sub)type of EB (Fine et al., 2000); (2) did not have any cognitive or psychological problems that could interfere with the study; (3) were fluent in the Dutch language; and (4) were willing and motivated to participate in the study (with informed consent). The definitive sample comprised eleven families with one or more children diagnosed with EB. In six families both the mother and father participated and in five families only the mother was interviewed. The children's ages ranged from two to nineteen years. Five of the children were diagnosed with EB simplex (EBS) Köbner type (EBS-K), Weber-Cockayne (EBS-WC), or Dowling-Meara (EBS-DM), two with junctional EB non-Herlitz type (JEB-nH), two with dominant dystrophic EB (DDEB), and two with mutilating recessive dystrophic EB (RDEB-mut). Two mothers and one father were also diagnosed with EB themselves (EBS-DM/DDEB).

Procedure

Prior to the acquisition of participants, the research was submitted to and approved by the local ethics committee. Thereafter the researcher informed the parents about the purpose and content of the study by means of an extensive letter. Participation was voluntary and parents who agreed to participate were asked to sign a written consent form. Two parents gave permission to use photographs of their child in this article. The same researcher completed all interviews. Parent interviews took place in their respective homes, to reduce the impact of time-consuming traveling. The interviews lasted from 1 to 1.5 hours. The interviewer also took notes on each child's condition, the interviewer-parent interaction, and the home environment to complement the recorded interview data (Ingham, Vanwesenbeeck, & Kirkland, 1999). During the interviews, paraphrasing and reflective listening were used to verify the understanding of the parents' reports. At the end of the interview, the parents were given the opportunity to summarize the most difficult problems that they experienced. The parents were then sent a copy of the transcribed interview, which gave them a chance to reflect on the interview report.

Data Analysis

The interviews were tape-recorded and transcribed verbatim. All data were analyzed in their original language to preserve the participants' original meanings. The transcribed interviews were subjected to content analysis with the qualitative software package Atlas-ti (Muhre & Friese, 2004). Atlas-ti offers tools to manage, extract, compare, explore, and reassemble meaningful pieces of text from large amounts of data in a systematic way. With the help of this software package, quotations that were relevant with regard to the aim of the study were coded based on meaning and judgments that the parents ascribed to their problems. The interview records were read carefully and then coded into small categories such as "time-consuming care," "uncertainty" and "restrictions in leisure time." These categories were further categorized into main themes, in the context of "care burden on the parent." The interview fragments were continuously compared to identify new themes and to specify the complex data in clear categories. Finally, the categories and main themes were described to a second researcher, new to the results of the study. Indistinctness in categories was discussed between the two researchers, resulting in the refinement of the themes and categories.

Results

It appeared that raising a child with EB involved divergent problems for parents, which affected many aspects of their lives. The most difficult problems the parents experienced can be divided into three main themes: (1) the burden of EB on their child's life, (2) the care burden on themselves and the family, and (3) the impact of EB on care providers. These three themes covered nine categories that seemed to best characterize parents' most difficult problems with EB.

Parents' Problems With the Burden of EB on Their Child's Life

Two categories, with respect to the burden of EB on their child's life, came to the fore as main problems that tormented the parents: (1) the child being different and (2) the child suffering pain.

The child being different: visible as well as invisible. EB is a skin disorder, which makes the skin of children with EB extremely fragile. Minor mechanical trauma such as hitting oneself, rubbing, scratching, or swallowing can result in blistering. Children with the most severe type of EB (RDEB) are so badly affected that they are covered with bandages from head to toe. Most of their fingers and toes also become fused because of scarring. As a result of skin contractures, their walking ability is to a great extent impaired, so most of the time they move around in an electric wheelchair. Children with the milder subtypes of EB simplex, junctional EB and dominant dystrophic EB, are less obviously affected. From time to time, their hands and faces show little blisters and wounds or scarring from blistering in the past. Children with junctional EB and dominant dystrophic EB are sometimes missing nails or have affected teeth. Some of these children's body parts are also protected with bandages. The skin disorder thus makes children with EB visibly different from healthy children (see Figures 1 and 2).

It was this visibility of EB that troubled parents of both mildly and severely affected children. For example, it made people stare at their child:

Lily was at the age kids start to crawl, so at that point her face was always battered. Then I checked whether people were staring and then, of course, they were. That drove me crazy, so I went looking for help. [mother of a mildly affected child]

Unkind remarks from others directed at the child were also troublesome for parents of both mildly and

Figure 1
A Mildly Affected Child With JEB-nH

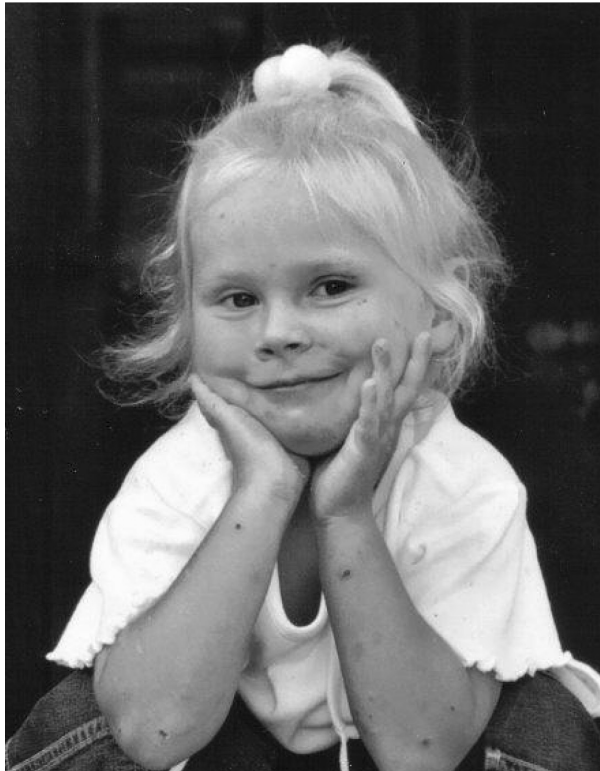


Figure 2
A Severely Affected Child With RDEB-mut



severely affected children. Some children were teased at school with funny names, or during swimming, when they got difficult questions like, “Is it contagious?” This could really bother the parents:

The saddest thing for me was when she came home from school, and there had been another confrontation, and someone had said something bad about it again. That was what I found the most difficult thing.
[mother of a mildly affected child]

In addition, the remarks were not only directed at the child, but the parents themselves also had to answer nasty questions. These were questions that brought out a lack of any notion about EB:

At first, when we walked with Lily through the village, people were inclined to say something about her, like, “Oh, how sad.” And there were people who asked questions like, “Has she fallen downstairs?” or “Have you been hitting her?” or “Does she have chicken-pox?” That kind of remark. Well, at first

they are very hard to deal with. [father of a mildly affected child]

Most parents acknowledged that this unpleasant behavior coming from others because of the visibility of EB was mainly a result of people in the immediate environment being unfamiliar with the illness. But there were also parents of mildly affected children who experienced a lack of understanding from others about the actual impact of EB on their child, just because the symptoms were mostly invisible. Their problem was that the people in the immediate environment could not see that their child with EB had limited possibilities in comparison to a healthy child. As a result, some friends or relatives expected that this child should be able to do more than he or she actually could, and accused the parents of being overly concerned:

When John is wearing his sweater and his trousers and all, then you don’t see anything. At the most all you see is that his hands are battered, that’s all. And

then people think that it is not that bad. They don't think about his feet, his knees, his elbows all being bandaged up at that same time. [mother of a mildly affected child]

Even the mother of an obviously severely affected child mentioned that she experienced a lack of understanding because of the "invisibility" of her child's illness. She mentioned that—because the affected skin of her child was totally protected with bandages—nobody, apart from the caregivers, could imagine the painful reality under that covering.

The child suffering pain: being in pain and causing pain. The more severely affected children are in pain during most of the day and night. This pain is caused by the various blisters and open wounds on the child's body. Mildly affected children suffer more often from occasional pain; for example, when they harm their skin surface while playing. Despite the differences in amount and occurrence of pain, parents of both mildly and severely affected children were afflicted by seeing their child in pain and by having few tools at their disposal to relieve that pain:

Of course, Lily doesn't like it [the pain], and I find that hard to deal with. (...) You don't always know the best way to help her. [mother of a mildly affected child]

But parents of both mildly and severely affected children were not just tormented by seeing their child being in pain. What affected them the most was that they had to cause their child pain; e.g., while taking care of the blisters and wounds:

I found it very hard to hurt my own child. You really have to shut yourself off completely. [parent of a mildly affected child]

Parents were also confronted with painful tasks when assisting the child in daily activities such as teaching their child to walk:

I would stand him up, but then he would start to cry really hard. With your first child, of course, you find that a terrible thing. You don't want to hurt him. [mother of a mildly affected child, about her child's first steps]

Parents of severely affected children, however, appeared to suffering from extra distress. This was related to the extensiveness and the duration of the painful tasks they had to do for the good of their child's

health. The extensive and time-consuming daily dressing changes, especially, were almost unbearable for the child and therefore for the parents. More often than not, the daily dressing changes resulted in a struggle with their child which was extremely hard to bear:

You know that you are going to be hurting your child for an hour or an hour and a half. That runs totally contrary to your feelings. You want to protect your child from this and make sure they don't have to go through it. [mother of a child with a severe subtype of EB]

There is always yelling and crying and screaming: "I don't want this and it hurts so much." Yes, that just stabs you in the heart. [mother of a child with RDEB]

Some parents therefore stressed the double role they were charged with. "I had no choice but to become a nurse, a doctor, and play all kinds of other roles" [mother of a severely affected child].

Parents' Problems With the Care Burden on Themselves and Their Family

Five categories were identified as putting a strain on the parents themselves and on the family. These are the uncertainty of the illness, restrictions on employment and leisure time, difficulties in organization of care, never being off-duty, and less time and attention for siblings and relationships.

Feelings of uncertainty. The most discussed and most troubling feeling expressed in the interviews was the sense of uncertainty. Parents of both mildly and severely affected children mentioned that, because of the unpredictability of their child's illness, one could never foresee when the child would have a bad day. This depended, for example, on the constant risk of their child falling, or on the low resistance or poor physical state of their child. This often resulted in restrictions on any spontaneous activities:

Just when you think things are all right, another blister develops. At the most unexpected times, really. Well, that's EB. [mother of a mildly affected child]

You never know what the day will bring. Sometimes we would be looking forward to a new day, and then he'd get blisters in his eye. He can fall, he can be in pain. He is often ill or on the verge of being ill. He has anemia, so he is always tired. [mother of a severely affected child]

In the interviews, parents of both mildly and severely affected children not only discussed the theme of uncertainty with respect to their short-term concerns, but also expressed feelings of uncertainty about the long-term prospects for their child's illness. Even when the subtype of their child's EB was clearly defined, some parents remained uncertain and fearful about the child's lifelong development: "You never know how bad it's going to be (. . .) You always carry that fear with you: will she get worse or will it just stay like this?" [mother of a mildly affected child]

For parents of severely affected children, the most difficult moments of uncertainty came when they were confronted with their child's condition worsening. Then questions such as, "How long do we still have," came up in their minds, and this thought sometimes even elicited the wish that "it was all over with," which immediately created a feeling of guilt about even thinking this way.

Restrictions in employment and leisure time. Parents also stressed that their previous ambitions and leisure interests had to take a back seat after their child with EB was born. Most couples decided that the care of the child would become the daily task of the mother, so that the father could keep his job and would be able to guarantee their income. Three mothers, two of a mildly affected child and one of a severely affected child, gave up their previous jobs because of their child's illness: "With the birth of Danny our world has turned upside down. I had to give up my job and since then I am at home full time." [mother of a severely affected child]

Furthermore, parents reported that the illness of their child also had a considerable impact on their social life. They mentioned that they had to give up a lot of their previous leisure interests, like holidays outside the country, going to the beach, or visiting relatives and friends frequently:

You could never make an appointment, and, if you did, nine times out of ten you had to cancel. Either James had just fallen, or something had happened so that he was completely exhausted and needed to sleep. And people just don't understand that; they think you worry too much. [mother of a mildly affected child]

Planning is actually very difficult. There are a lot of things that you would like to do, which you can't. We can't go to the beach. The wind hurts him. When it's too hot, you can't go out, when it's too cold, you

can't go out, because either way his skin breaks open. You are very restricted. [mother of a severely affected child]

Difficulties in organization of care. It should be mentioned that in most families it was the father who managed the organizational aspects of care, including the special requests such as a hoist for bathing, special transportation to the child's school, or financial support for dressings and gauze. From their remarks, it became clear that they experienced the organizational aspects of care as a complex and time-consuming task. In other words, for almost every request, they had to apply to a different agency or authority: "Every time there was so much to arrange that you actually needed a secretary for a day and a half to get everything set up." [father of a mildly affected child]

Severely affected children, for instance, need an enormous amount of bandages, gauze, and ointments. Some of their mothers described being worried about bandages and ointments not being delivered in time, so that they were afraid they might run out of supplies for wound care. In this context, parents of both mildly and severely affected children mentioned the wish to have a care manager. In their eyes, that had to be someone who could organize all medical, financial, and administrative aspects of care and arrange the multiple appointments with the hospital, the rehabilitation center, the dentist, and other professional services. According to the parents, this would give them more leisure time and energy to spend with their affected child.

Never being off-duty. One of the first messages from parents of severely affected children during the interviews was that they experienced the care of their child as a rather time-consuming task. One mother stated that this was merely because of changing the dressings of her child every day. For her, this could take more than 2.5 hours, depending on the amount of professional assistance she could get. The remainder of the time she was occupied with preparing medicines and food, stretching and splinting the hands to prevent the fusion of fingers, as well as helping with daily activities for which her child gradually became dependent on her. As she said, "I can't leave Alex alone for longer than half an hour. He can't go to the toilet himself, he can't pour himself a drink. You're always worried that he will fall or that he will hurt himself." Another couple mentioned that they had the physically exhausting task of continuously assisting their child in toileting, playing games, blowing his nose, or eating a meal. But parents of mildly affected

children also mentioned that the illness of their child could take up much of their time. In their case, the burden was not the physical pain of dressing changes and the dependency of the child, but instead was the nuisance of traveling around the country for all the various kinds of treatments their child needed, which was experienced as a time-consuming job.

Care for the child was not just physically stressful for some parents; that is, parents of both mildly and severely affected children also experienced care for their child as a mental burden—times two. For the first parent, this was caused by constant worry about all aspects of the illness. Said the mother, “It just exhausts you. You live with it twenty four hours a day. It is different from being at work; you can’t close the door behind you and walk away. You cannot escape it. It’s always on your mind.” For the other parent, the mental burden was a result of difficult treatment-related choices that had to be made. These were choices that were not for improvement, but instead were because the child was on the decline. For example, there was the difficult choice of whether or not to start using a wheelchair, whether or not to operate on the hands again because of the constant webbing of the fingers, or whether or not to fit a percutaneous endoscopic gastrostomy tube because of the child’s problems with swallowing, eating, and the resulting malnutrition and growth retardation. A mother about weighing one choice against the other, said, “You always have to choose the lesser of two evils. You can never say that if you do this, then you solve that and that. It is always palliative, with several other problems appearing afterwards. Actually, it’s just never-ending.”

Family problems: feelings of guilt and growing apart. As a result of this time-consuming care most parents of severely affected children mentioned that they found it rather difficult to divide their attention equally among all family members. One mother, for example, found it really hard as she noted that her healthy daughter had to amuse herself for all those hours when her brother’s dressings were being changed: “I often feel guilty about that. Then I think that she’s just going to have to wait her turn. Always.” Feelings of guilt about the unequal division of their attention seemed to increase the burden of care.

Most of the parents of both mildly and severely affected children experienced the illness of their child as also having an impact on their relationship. In four of the eleven families interviewed the parents were divorced. Two of the divorced mothers emphasized

that their separation was to a certain extent because of the illness of their child. In addition to that, a number of non-separated mothers also mentioned that they sometimes had the feeling of being on their own or having grown apart from their husbands because of the care needed for their child. One of the causes of this feeling seemed to be a result of the dissimilarity in the way the parents dealt with the illness. For example, some parents experienced a dissimilar need to talk about their concerns or they had disagreements about whether or not to hand care over to professionals. In addition, not being able to pay enough attention to the relationship or not having enough leisure time together also caused a feeling of growing apart: “When we sit down in the evening, we only talk about our worries and about the things that have to be done.” [mother of a severely affected child]

Parents’ Problems With the Impact on the Care Providers

Two categories could be identified that were related to difficulties parents had with care providers. These were the ignorance and resulting lack of skills of the care providers, and their resistance to the difficult aspects of care.

Ignorance and lack of skills. Most of the parents encountered several problems because of unfamiliarity of professional health care providers with EB. When seeing their child at birth, covered with blisters or missing parts of their skin, parents were of course very eager to know what was wrong. But their experience was that nobody in the hospital could tell them:

In the hospital, nobody knew what EB was. Can you imagine, in a hospital? They had never heard of it. Not many hospitals seem to know much about it. [mother of a mildly affected child]

After finding out what the exact diagnosis was from a dermatologist familiar with EB, parents still encountered several obstacles because of the ignorance of professional health care providers regarding EB. They mentioned, for instance, that some dermatologists gave an incorrect description of the progress of the illness, or that nurses did not use the right materials for the blisters, such as using sticking bandages, which further damaged the skin.

Some parents described the lack of knowledge about EB as being even worse when it came to home care providers. As a result, the parents of both mildly

Table 1
Parents' Nine Most Difficult Problems in Caring for a Child with EB

| Concern | Example |
|--|--|
| The visibility of the illness | "People were inclined to say something about her." |
| The invisibility of the illness | "People think that it is not that bad." |
| Parents "causing" pain | "I found it very hard that you ended up hurting your own child." |
| Uncertainty: long-term | "You don't know how bad it will get." |
| Uncertainty: short-term | "You never know what the day will bring." |
| Restrictions on employment and leisure time | "Planning is actually very difficult." |
| Difficulties in the organization of care | "You actually needed an administrative employee." |
| Never being off-duty | "I can't leave him alone longer than half an hour." |
| Reduced attention for siblings | "I feel rather guilty about that." |
| Problems with the relationship | "We only talk about our concerns." |
| Ignorance and lack of skills of care providers | "In the hospital, nobody knew what EB was." |
| Resistance to difficult care | "It just stabs them [the home care providers] in the heart." |

and severely affected children found themselves confronted with unskilled care providers who lacked the specific expertise that the parents had developed during daily treatment of their child:

At a certain point we were able to get home care. That meant that every three days there was a different nurse on the doorstep, who we had to show how to do John's dressings. (. . .) And when they started, you thought "I could have done that four times faster. Then John could go back to sleep." That cost us so much more time and energy than it gave us. So, then you just stop. [mother of a mildly affected child]

In many cases, the parents felt that they had to discover all by themselves how to handle this little-known skin disorder. Mothers of both mildly and severely affected children expressed the fact that they felt they were the experts on EB instead of the professional health care providers. This made them feel as if responsibility for the quality of care was all on their shoulders. Some of them described this as "the world [being] upside down."

Resistance to difficult care. Another problem with home care, exclusively for mothers of severely affected children, was that their care providers found it very hard being confronted with the painful struggle with the child every day when helping change the child's dressings. They mentioned that some of the skilled home care providers simply didn't want to do this stressful kind of care work more than once a week, or that after a while they just gave up: "It just stabs them in the

heart. That's why they don't actually want to do it [the care] more than once or twice a week. So, I end up with a lot of different ones [care providers]." As a result, these parents were frequently searching for stable care providers, who might have some endurance.

Despite the impact of the child's illness on the parents' paid employment and leisure time, most of the parents of mildly affected children stayed away from the help of home care providers. Most of these parents wanted to perform the care themselves. Although parents of severely affected children usually did have home care, they also tried to keep a tight control over the care given. This surprising decision seemed to be a result of their experience with the ignorance and lack of skills of professional healthcare and home care providers, and the feeling that they themselves were the experts in care.

Discussion

There are a small number of studies to date that address the difficulties of parents of a child with EB (Landsdown et al., 1986; Lin & Caldwell-Brown, 1996; Fine, Johnson et al., 2005). Aspects of these studies that are in line with our study include parents' difficulties with staring and difficult remarks made to the child, unexpected life changes, time-consuming care, relationship problems, and the ignorance of care providers. In contrast to the rare skin disorder EB, parental experiences with more regular childhood chronic illnesses, such as cancer, cystic fibrosis, congenital heart disease,

spina bifida, or diabetes are far more frequently discussed in the literature (e.g., Cohen, 1995; Diehl, Moffitt, & Wade, 1991; Fisher, 2001; Hewitt-Taylor, 2005; Ievers, & Drotar, 1996; Johnson, 2000; Monsen, 1999; Ratcliffe, Harrigan, Haley, Tse, & Olson, 2002). In the following sections, we will therefore bring out the similarities and differences between previous EB research and other chronic childhood illnesses, including another chronic skin disorder, atopic dermatitis. This is a more common skin disease of early childhood, with itchy and dry skin combined with eczema lesions as its main features.

The Child Being Invisibly Different

Lack of understanding from others tormented parents of children with all types of EB. This lack of understanding manifested itself in teasing and staring at their child, as well as in nasty remarks and unpleasant questions to both the child and the parent. Sixty percent of children with atopic dermatitis also experienced teasing and bullying, resulting in psychological problems of the parents (Lawson, Lewis-Jones, Finlay, Reid & Owens, 1998).

In contrast to the well-recognized problematic visible aspects of EB (Landsdown et al., 1986), we found that parents were also bothered by lack of understanding from others because of the invisible aspects of the child's illness. In cases where the blisters, wounds, and scars were covered with clothes and bandages, other people tended to underestimate the impact of the illness on their child's well-being.

Parents Causing Pain to the Child

The pain that the parents themselves had to cause their child to protect the skin and to ensure necessary daily activities tormented the parents especially, as it often resulted in a struggle between the child and the parent. This struggle was most bothersome for parents with a severely affected child because of the duration and extent of the painful tasks that needed to be done almost every day. However, this problem was not specific only for children with severe EB. Parents of children with chronic atopic dermatitis also mentioned a struggle with their child, because of the child's overwhelming need to scratch during undressing and the painful sticking of eczematous lesions to the skin (Elliot & Luker, 1997). Nevertheless, we believe that the duration and amplitude of the pain in severe EB is of a higher order compared to that in chronic atopic dermatitis.

Landsdown and colleagues (1986) already found that more than half of parents of a child with EB mentioned difficulties with dressing changes. He mentioned that the major difficulty parents experience was in getting bandages off without the bandages sticking and without removing skin. We discovered that parents' problems with dressing changes were more than just a practical problem. Difficulties with dressing changes were also related to the difficult emotional task of the parents, which involved hurting their own child during treatment.

The parents found it very hard to combine the pain-inflicting professional care required of them with their desire, as a dedicated parent, to protect their child from pain. The only way to deal with this inner conflict was to try to separate themselves from their emotions during the painful tasks they had to perform for the good of their child's health. In a review study, Ratcliffe et al. (2002) also found that parents' role conflict was a core theme for parents of children with other chronic illnesses. They described this as the parents being in an ongoing flux between being a parent and being a care provider. However, it seems that EB is the first disease for which parents have the difficult task of causing intense pain to their child for the sake of the child's health. This treatment can be so painful for the child that it becomes one of the main care burdens for the parents.

The Unpredictability of the Illness

Parents of children with all kinds of EB were forced to confront the short-term as well as the long-term unpredictability of their child's skin disease. For parents of severely affected children, feelings of uncertainty were all the more intense, as they incorporated serious concerns about the diminished life expectancy of their child because of the progressive character of the illness. For parents of children with all types of EB the unpredictability of the illness seemed to result in difficult life changes, including restrictions in working abilities, leisure time, and social contacts. Lawson et al. (1998) also found that almost half of the parents of a child with atopic dermatitis experienced restrictions in going out and pursuing hobbies because of the exhausting character of the child's disease.

Several studies indicate that caring for a chronically ill child involves constant uncertainty, although most of the time this is at a subconscious level (Cohen, 1993, 1995; Diehl et al., 1991; Fisher, 2001; Monsen, 1999). In a study on parental uncertainty, Cohen (1995) identified seven triggers that had the

potential to cause a heightened level of uncertainty for parents of a chronically ill child, including (1) routine medical appointments, (2) body variability, (3) key words and provocative questions, (4) changes in therapeutic regime, (5) evidence of negative outcomes, (6) new developmental demands, and (7) nighttime. In our study, we discovered that parents of a child with EB experienced minor symptoms or variations from the child's norm (body variability) and evidence of aggravations and the child going downhill (negative outcomes) as being the greatest triggers of uncertainty.

Difficulties With Organization of Care

Most parents of children with all kinds of EB experienced the managing of the organization of care as an additionally demanding task. In other words, the fragmentation of care facilities, which turned applications for all kinds of medical needs into a time-consuming task, was what bothered them the most. For parents of severely affected children, the probable absence of prerequisites for delivering adequate home care could specifically contribute to additional parental stress. In a focus group study, Diehl et al. (1991) also identified the fact that children with severe chronic conditions require an extraordinary quantity and quality of care, which demands complex orchestration and which underscores the need for a competent care manager.

The Mental Burden of the Illness

Parents of severely affected children seemed to experience both a physical and a mental burden as a result of their child's illness. The continuous-care burden seemed to trouble them to a great extent and contributed to a feeling of never being off-duty. Parents of mildly affected children were only occasionally confronted with time-consuming care, so they were able to have some time off. However, for some of these parents, the illness of their child still occupied their minds twenty four hours a day. Other studies also found that the sense of never being off was the most frequently reported burden of parents of seriously chronically ill children (Ratcliffe et al., 2002) and that parents had real problems with the arduous day-to-day treatment of their child's illness (Ievers & Drotar, 1996). We discovered from our study that the sense of never being off-duty also included a mental burden for parents, including almost permanent worrying and having to make difficult choices in treatment which were a sign that their child was on the decline.

Family Problems

It seems that the intensity of care needed for children with EB impacts all family members. Feelings of guilt about the reduced attention for siblings and their feelings of growing apart in the relationship troubled many parents. The little time and attention left for siblings and the relationship created a situation in which, in some cases, the balance of the family was disturbed, and some couples ultimately decided to divorce because of this. It was quite a source of concern that, although some parents of a child with EB were not separated, the mothers often still had the feeling of facing it alone or growing apart because of caring for their child. Other studies also identified the fact that a child with a chronic (skin) disorder can cause minor as well as major disruptions in intrafamilial relationships (Fine et al., 2005; Diehl et al., 1991; Ievers & Drotar, 1996; Lawson et al., 1998). Problems that put strain on the parents' relationship included, for example, rarely doing things as a couple, difficulties in not talking about the affected child all the time, being overprotective, or lack of energy left for activities as a couple.

Ignorance of Care Providers

Lin and Caldwell-Brown (1996) has stressed the problem that there are dermatologists who might never see a case of EB in a lifetime of practice, and that many physicians have never even heard of it. For these reasons, parents might feel bewildered and frustrated when these professionals are required to support them in their uncertainty and their search for adequate care for their child. The resulting wish of parents to maintain control over care, which the parents discussed in our study, was also found in another review study by Ratcliffe et al. (2002). When parents frequently experience unreliable and unskilled care, they naturally develop the need to be continuously supervising or teaching the care professional.

Conclusions

Epidermolysis bullosa is not a single disease; it consists of at least twenty-five different subtypes with varying degrees of severity. The illness can range from mild forms with only local involvement of the skin surface to severe forms involving various organs entailing chronic progression. The aim of this study was to explore the main problems of parents of a child with EB and the probable differences in experience between parents of mildly and severely affected

children. Despite the great variance in the clinical picture of the types and subtypes of EB in children, generally most problems parents of children with EB have appeared to be quite similar. Parents of children with all types of EB experience problems with the (in)visibility of the illness, the pain of the child, uncertainty, restrictions in employment and leisure time, never being off-duty, problems within their own relationship, and problems with the ignorance and lack of skills of care providers. However, when clearly specifying these problems, they did appear to differ in intensity and gravity; that is, parents of severely affected children have to deal with more extensive, time-consuming, and painful care and more intense feelings of uncertainty and of never being off, along with feelings of guilt about the unequal attention given to other family members.

Future Directions

Parents caring for a chronically ill child appear to experience disease-specific as well as generic problems. From our study, it became clear that more generic problems of parents of a child with a chronic skin disease include problems with the teasing of the child, the daily struggle with the child during treatment, problems with interfamilial relationships and restrictions in work and leisure time. Intervention programs for parents of a child with a chronic skin condition should therefore include elements which aim to (1) improve parental coping with a visibly affected child, (2) strengthen family relationships, and (3) create opportunities for parents to have some respite.

In contrast to other chronic childhood skin disorders, specific problems for parents of a child with EB are the diminished life expectancy of certain subtypes, the time-consuming and extremely painful care, and the ignorance of the disease on the part of the healthcare professionals because of the rare incidence of EB. Although there is a great deal of support available for parents of a child with EB in the Netherlands (a home care program, an expert center in the medical center and a patient organization), parents still explicitly express the need for a competent care manager. Parents' difficulties with performing painful treatments, their problems with the lack of expertise of many home care providers, and their problems with the fragmentation of care facilities all indicate that there is a great need for a competent and expert care manager in the home situation. This care manager should be able to adequately answer questions families have about all aspects of EB, and he or she should be well-informed about the care facilities

available for these parents and their children. The care manager should be able to indicate whether the support of a professional is needed, and what kind of support is most wanted. Is there a need for psychological help, or more practical support—or maybe both? Parents should not have the feeling they have been left all alone in caring for their child with EB.

Appendix Abbreviations

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| EB | epidermolysis bullosa |
| EBS | epidermolysis bullosa simplex |
| EBS-DM | epidermolysis bullosa simplex, Dowling-Meara type |
| EBS-K | epidermolysis bullosa simplex, Köbner type |
| EBS-WC | epidermolysis bullosa simplex, Weber-Cockayne type |
| JEB | junctional epidermolysis bullosa |
| JEB-nH | junctional epidermolysis bullosa, non-Herlitz type |
| DDEB | dominant dystrophic epidermolysis bullosa |
| RDEB | recessive dystrophic epidermolysis bullosa |
| RDEB-mut | recessive dystrophic epidermolysis bullosa, mutilating |

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